Anomalous left coronary artery from the right pulmonary artery with aortic fusion

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Anomalous left coronary artery from the right pulmonary artery (RPA) with aortic fusion is a rare variant of anomalous left coronary artery from the pulmonary artery (ALCAPA). Three consecutive cases of this ALCAPA variant were managed, including a case with associated pink tetralogy. The challenges that can present in the diagnosis and treatment of this rare condition are described.

CLINICAL SUMMARY

A 3-month-old male infant was referred for elective repair of tetralogy of Fallot (TOF) with pulmonary stenosis. Echocardiography was consistent with a clinical picture of a “pink” TOF with normal coronaries. A total subannular repair of TOF with pulmonary stenosis was performed on cardiopulmonary bypass. During weaning, high left atrial pressures were noted with depressed left ventricular function. A transesophageal echocardiogram demonstrated satisfactory anatomic repair. Coronary anatomy was specifically examined. The left coronary artery appeared to originate from the left coronary sinus by both echocardiography and direct inspection. The patient was placed on extracorporeal membrane oxygenation (ECMO). Catheterization on ECMO the next day demonstrated an anomalous left main coronary artery (LMCA) arising from the RPA (Figure 1). At reoperation, careful dissection of the LMCA confirmed its origin from the RPA very close to the aorta. The ostium of the LMCA was excised as a button, but the vessel 3 or 4 mm distally could not be separated from the aortic wall to which it was fused. No intramural segment could be detected through an aortic incision and by probing the coronary artery. The short length of the coronary artery between the RPA and the point of fusion to the aorta did not allow direct transfer of the coronary button without risk of kinking. Thus, it was sutured to the lateral edge of an opening made in the ascending aorta with pericardium used to roof the opening (Figure 1). A femoral vein homograft segment was interposed between the distal pulmonary

bifurcation and the proximal divided main pulmonary artery to prevent tension over the reimplanted coronary artery. Postoperative transesophageal echocardiogram showed good flow in the LMCA. The patient was weaned from ECMO 5 days later. Follow-up catheterization after 1 month revealed obstruction of the LMCA at the site of aortic anastomosis. This may be a consequence of late revascularization or kinking. The patient had a complicated and extended hospitalization, but he was eventually discharged. He died suddenly at home a few months later.

The same condition was encountered at Children’s National Medical Center in 2 other male infants aged 4 and 6 months over a 6-month period. At operation, the left coronary artery had the same aortic fusion with RPA origin as previously mentioned. There was no evidence of dimpling within the left coronary sinus to attempt any fenestration. Given the short extramural course before fusion with the aorta, we transected the LMCA at its pulmonary end, which was oversewn. The LMCA was then filleted open to its aortic attachment, where a small intramural segment measuring less than 0.5 mm was observed. The filleted left coronary artery was directly anastomosed to the leftward

FIGURE 1. Coronary transfer technique used in patient 1, who had aortic fusion but no intramural segment. The short segment between the RPA and the point of aortic fusion is at risk of kinking. LPA, Left pulmonary artery; MPA, main pulmonary artery; RPA, right pulmonary artery.

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TABLE 1. Previous reports of anomalous left coronary artery from the right pulmonary artery with aortic fusion

	Atik and colleagues ² N = 4	Turley and colleagues ³ N = 1	Goldberg and colleagues ⁴ N = 3	Adachi and colleagues ⁵ N = 1	Our series N = 3
Origin	RPA	RPA	RPA	RPA	RPA
Associated lesions	None	None	Restrictive VSD	None	TOF
Preoperative diagnosis					
ALCAPA	+	+	+	+	+(2/3)
Anomalous course	–	–	–	+	–
Extramural course	?	?	Short	Short	Short
Intramural course	+(3/4)	+	+(small)	+	+(small) Absent in 1
Surgical correction	Unroofing alone in 3 Coronary reimplantation without Unroofing in 1	Unroofing alone	Unroofing with direct reimplantation in 2 Coronary reimplantation alone without unroofing in 1 → Revision of anastomosis with unroofing	Unroofing alone	Unroofing with direct reimplantation in 2 Coronary reimplantation with pericardial roof without any unroofing
Results	3 survived 1 death in reimplantation alone	Survived	3 survived	Survived	2 survived 1 death in reimplantation alone

VSD, Ventricular septal defect.

and posterior aspect of the aorta. Postprocedure transesophageal echocardiogram in both patients demonstrated good flow in the LMCA. Both these patients did well in the postoperative period. Echocardiography at follow-up more than 1 year later revealed significant improvement in left ventricular systolic function and antegrade flow in the LMCA.

DISCUSSION

Anomalous left coronary artery from the RPA with aortic trajectory and fusion was first described by Atik and colleagues¹ in 1988. Because there was also a well-defined intramural course in Atik and colleagues' cases and treatment was easily accomplished by fenestration, they termed this "a difficult condition to diagnose but easy to correct."

A thorough review of the literature reveals only 9 additional previous reports of such a variant²⁻⁵ (Table 1). Although the diagnosis of ALCAPA was made in 11 of the 12 cases, the peculiar aortic trajectory of this variant was reliably predicted in only 1 case.⁵ The close approximation of the LMCA to the left aortic sinus, its vertical course running parallel to the ascending aorta, and the drop-out of the thin wall separating it from the aorta make preoperative characterization of the trajectory difficult by echocardiography.^{2,6} Barbero-Marcial and colleagues⁶ defined the angiographic criteria for the diagnosis of this condition. In the setting of a significant left to right shunt and elevated pulmonary artery pressures, as was the case in our first patient, the diagnosis becomes even more difficult because there is little or no reversal of flow in the left coronary artery. At operation too, it is as difficult to diagnose this

condition by inspection because the LMCA appears to arise normally from the left sinus of the aorta. A preoperative magnetic resonance angiography would be useful in the diagnosis of this rare condition whenever it is suspected. In all previous reports, surgical correction was achieved by unroofing the intramural segment alone in patients with a sufficient length of vessel that was intramural to allow this (Table 1). Patients who underwent unroofing alone or as part of a procedure in addition to direct reimplantation survived. Both the deaths reported previously occurred in patients who had coronary transfer alone. The principle to bear in mind at operation is that the coronary artery should not be allowed to kink during reimplantation. If there is a well-defined intramural segment, then unroofing is easy and the best option. If the intramural segment is small, then unroofing the small segment, filleting the coronary artery, and anastomosing to the aorta as described in our other 2 patients could be used with good results.

CONCLUSIONS

Anomalous left coronary artery from the RPA with aortic fusion is difficult to diagnose and may be challenging to repair when there is minimal intramural course.

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Placement of 2 implantable centrifugal pumps to serve as a total artificial heart after cardiectomy

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In recent years, the role of ventricular assist devices for management of chronic heart failure has continued to increase. This increase stems mainly from the success of improved technology, greater surgical experience, and increasing experience with management of ventricular assist device–related complications. Collectively, these advances have led to overall better outcomes.

The HeartWare Ventricular Assist Device (HVAD; HeartWare International, Inc, Framingham, Mass) has become an established mechanical support therapy for left ventricular failure.^{1,2} Moreover, the use of the HVAD for biventricular support has been successfully reported.^{3,4}

We report the case of a 51-year-old man presenting with a ventricular septal defect after myocardial infarction, not amenable to surgical repair. Two centrifugal blood pumps were then implanted after cardiectomy, serving as a total artificial heart intended to be used as a bridge-to-transplant device.

CLINICAL SUMMARY

The patient was a 51-year-old man who initially came to a regional hospital with acute coronary syndrome. The

cardiac catheterization performed showed complete occlusion of the right coronary artery proximally, resulting in a large posterolateral and septal myocardial infarction. On echocardiography, biventricular failure and a large ventricular septal defect were seen. Despite maximal inotropic support, a LifeBridge extracorporeal support system (LIFE-BRIDGE Medizintechnik AG, Ampfing, Germany) was implanted for hemodynamic stability. The patient was transferred by helicopter on an emergency basis to our institution in Hannover, Germany.

Transesophageal echocardiography was repeated, confirming biventricular failure, a large ventricular septal defect, and new-onset aortic regurgitation. The aortic regurgitation limited the effectiveness of the life support system, thereby mandating surgical intervention.

Intraoperatively, after median sternotomy the patient was switched from the life-support system to a conventional heart–lung machine with transfemoral venous cannulation and arterial cannulation of the ascending aorta. Initially, aortic valve replacement with a biologic prosthesis was performed. On visual inspection after ventriculotomy, however, a massive posterior infarction with destruction of the posterolateral wall was seen. This included the entire ventricular septum and anterior papillary muscle, far beyond conventional surgical repair.

The only option was the implantation of a total artificial heart. Under complete cardiopulmonary bypass, the aortic valve prosthesis, the pulmonary valve, and both ventricles were resected so as to preserve the atria and the annuli of both atrioventricular valves. An epicardial HVAD sewing ring was implanted on the ventricular aspect of the mitral annulus with a 2-0 running Prolene suture (Ethicon, Inc, Somerville, NJ). The suture was supported by felt strips running outside the annulus around the left atrium. The sewing ring and the

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